Functional Tumors of the Organ of Zuckerkandl

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Chromaffin-reacting pheochromocytomas of the adrenal medulla are the most frequently encountered functional paraganglionic neoplasms. However, extra-adrenal pheochromocytomas as well as non-chromaffin paragangliomas, including those of the carotid body and glomus jugulare, may produce symptoms from catecholamine secretion. One of the extra-adrenal sites from which these tumors arise is from a collection of para-aortic, paraganglion cells around the origin of the inferior mesenteric artery. This collection of paraganglia was described in fetuses by Zuckerkandl[59] in 1901 and has subsequently been referred to as the organ of Zuckerkandl. The diagnosis and management of these neoplasms differ somewhat from that of adrenal pheochromocytomas, but excellent results are often obtained by excision of these lesions. Four patients with functional tumors of the organ of Zuckerkandl are reviewed together with the other reported cases in the literature.

SINCE 1961 4 instances of functional tumors of the organ of Zuckerkandl have been treated at The New York Hospital-Cornell Medical Center (NYH-CMC). We have found only 46 other instances of organ of Zuckerkandl neoplasms, which can be considered in 3 groups: those discovered at autopsy, those treated surgically by removal of the tumor and those in which operation was embarked upon but the tumor was not removed (Tables 1 to 3).

Group I: Tumors discovered at autopsy. Of 14 tumors, 8 in males and 6 in females, the age range was 15 to 81 years with a median of 53 years. Three tumors (21%) were considered malignant; the remaining 11 (79%) were classified as benign.

Group II: Tumors removed at operation. This is the largest group, 30 in all, 18 males and 12 females. The ages ranged from 15 to 55 years with a median of 32 years. Four (13%) of the tumors were considered to be malignant and 26 (87%) were benign.

Group III: Tumors found at surgery but not removed. There were 6 cases, 2 males, 3 females, 1 not identified. The ages were from 10–60 years with a median of 26 years. Attempted excision was not completed in 2 patients because of intraoperative death. In one case only a biopsy was done and in 3 the tumor was observed and considered inoperable. One of the latter 3 received irradiation and died 11 years later.

The data included in many of the case reports leave much to be desired. The symptoms which led to consideration that these patients had such a lesion are often poorly described and there is paucity of evidence to establish how a diagnosis was made preoperatively. In particular, there is inadequate documentation of whether there was excessive catecholamine secretion by the tumors. Furthermore, followup was not given in 18 of the 30 patients in whom operation was presumably successful. Of the remaining 12, 2 were well 12 years later, 8 were alive 3 years or less after operation and 2 were dead. Thus there is great need for case reporting of those who have a tumor in the organ of Zuckerkandl discovered at autopsy or operation.

Case Reports

Case 1. In 1961, a 17-year-old boy (NYH 89 79 71), previously believed to be well, developed headache and within a few hours collapsed and then had a grand mal seizure. The first physician to see him found a blood pressure of 180/115. At rest after admission to the hospital, the blood pressure was 150/110. Laboratory studies were essentially within normal limits except for total urinary catecholamine excretion which was 2422 μg/24 hr (normal total catecholamines, 200–250 μg/24 hr). A diagnosis of pheochromocytoma was made. The abdomen was explored and a lobulated retroperitoneal tumor was found over the inferior vena cava and abdominal aorta. The vascular supply was by several small arteries, arising directly from the aorta. During manipulation of the tumor, the pulse increased from 110 to 138 and the blood pressure from 140/90 to 150/100. The lesion was removed intact along with 2 adherent lymph nodes. The adrenal glands were uninvolved by tumors. The patient made a satisfactory recovery and is reported to be well 14 years later.

Case 2. A 39-year-old mother of 3 children (NYN 90 12 57) was admitted in 1962 for evaluation of headaches. Her blood pressure was 190/110 and was known to have been elevated for 16 years. Initially, this had been assumed to be related to a toxemia of

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TABLE 1. *Tumors of Organs of Zuckerkandl Revealed at Autopsy*

Author	Patient		Diagnosis
	Age	Sex	
Hausmann[21]	53	M	Benign
Handschin[19]	45	M	Benign
Nordmann[44]	53	M	Benign
Reichardt[49]	53	F	Benign
Cragg[6]	39	F	Benign
Gellerstedt[13]	38	M	Benign
Podloucky[48]	51	M	Benign
Fingerland[9]	71	M	Benign
McCullagh[37]	28	M	Benign
Koster[33]	34	F	Benign
Ortega[46]	81	F	Malignant
Isaacson[28]	15	F	Malignant
Moir[42]	55	F	Benign
Melicow[38]	56	M	Malignant

pregnancy, but urinary catecholamine excretion was high, 332.5 μg/24 hr. Because the adrenal arteries were normal on arteriogram, it was concluded that tumor was extra-adrenal, and at operation, through a long midline incision, both adrenals appeared normal but a hard, nodular tumor lay between the aorta and the inferior vena cava. As the operation was being completed after resection of the tumor, the patient became cyanotic and hypotensive and died despite vigorous resuscitative efforts.

Case 3. In 1966 a 15-year-old boy was found to have an abdominal tumor at the level of the umbilicus. He had been seen as an outpatient over a 4-year period for a variety of complaints, including visual disturbances, dermatitis, minor traumatic injuries, headaches and precardial pain during exercise. A diagnosis was not established; blood pressures were 144/86 and 132/80 at the time the mass was found, but 6 days later were recorded as 200/100. Total urinary catecholamine determination was 1560 μg/24 hr. During induction of anesthesia, the blood pressure increased to 220/160. A retroperitoneal para-aortic mass was found and excised. After clamping the veins, the patient became hypotensive and required intravenous levarterenol for 24 hours. The adrenals appeared normal grossly. The postoperative course was satisfactory with normotension for 3 years, after which followup attempts were unsuccessful.

Case 4. A 21-year-old man (NYH 137 11 02) was known to have "high blood pressure" for 10 years. In 1974 he was explored at another hospital for a lower abdominal retroperitoneal mass which was not removed because of its vascularity and profuse bleeding. The biopsy report was "pheochromocytoma." He was then admitted to The New York Hospital where an aortogram and inferior mesenteric arteriogram showed a vascular tumor extending from just below the right renal artery to the level of the hypogastric artery and extending across the midline, a short distance beyond the left side of the aorta (Fig. 1). The inferior mesenteric artery was displaced by the mass and supplied branches to it. Total urinary catecholamine excretion was 264 μg/24 hr. At exploratory laparotomy numerous vessels supplied the tumor from the aorta and drained into the vena cava. These were divided individually so that the tumor could be raised from its bed. Because of the difficulty in handling the numerous vessels, the operation required several hours of meticulous dissection and numerous transfusions. The postoperative course was uneventful and the patient was free of complaints when last seen 18 months after surgery.

Pathological Findings

The tumors were all lobulated, fleshy masses (Fig. 2). A definitive fibrous capsule was evident in only one tumor. The color of the cut surfaces was light brown or reddish-brown. Prominent blood vessels and hemorrhage were seen in 2 cases and cystic degeneration in one. The recorded weights ranged from 78 to 244 grams and the greatest dimension was 10 cm.

Microscopically, all of the tumors had a population of polygonal cells with fairly uniform, round to oval nuclei and a faintly, slightly granular, indistinct, eosinophilic cytoplasm which tended to be arranged in alveolar nests surrounded by a richly vascular connective tissue stroma

TABLE 2. *Surgical Excision of Tumor*

Author	Patient		Diagnosis	Followup
	Age	Sex		
Strangl[55]	32	M	Benign	—
Bauer[1]	40	M	Benign	—
Fullerton[12]	27	M	Benign	Well, 2 yr postop
Richardson[50]	28	M	Benign	—
Molinatti[43]	24	F	Benign	—
Forssel[10]	38	F	Benign	Recurrence, 12 yr postop
Cook[4]	31	M	Malignant	—
Cook[4]	42	F	Benign	—
Cook[4]	37	M	Benign	—
Heubner[24]	52	M	Benign	—
Holsti[26]	53	M	Malignant	Died, 10 mon postop
Holsti[26]	24	F	Malignant	Improved, 1 mon postop
Mintiens[40]	20	M	Benign	Well, 6 mon postop
Van Zyl[58]	31	F	Benign	—
Joseph[30]	33	M	Malignant	Metastasis, 33 mon postop
Lulu[34]	32	M	Benign	—
Hillestad[25]	—	F	Benign	—
Cornog[5]	27	M	Benign	Well, 1 yr postop
Malter[35]	55	M	Benign	—
Hendry[23]	52	F	Benign	—
Kinkhabwala[32]	54	F	Benign	—
Kinkhabwala[32]	31	F	Benign	—
Czekala[7]	43	F	Benign	—
Hahn[18]	19	M	Benign	—
Deoreo[8]	16	M	Benign	—
Shepard[52]	28	F	Benign	Well, 7 mon postop
NYH-CMC	17	M	Benign	Well, 12 yr postop
NYH-CMC	39	F	Benign	Death at operation
NYH-CMC	15	M	Benign	Well, 3 yr postop
NYH-CMC	21	M	Benign	Well, 1 yr postop

TABLE 3. *Attempted Surgical Excision (Abandoned)*

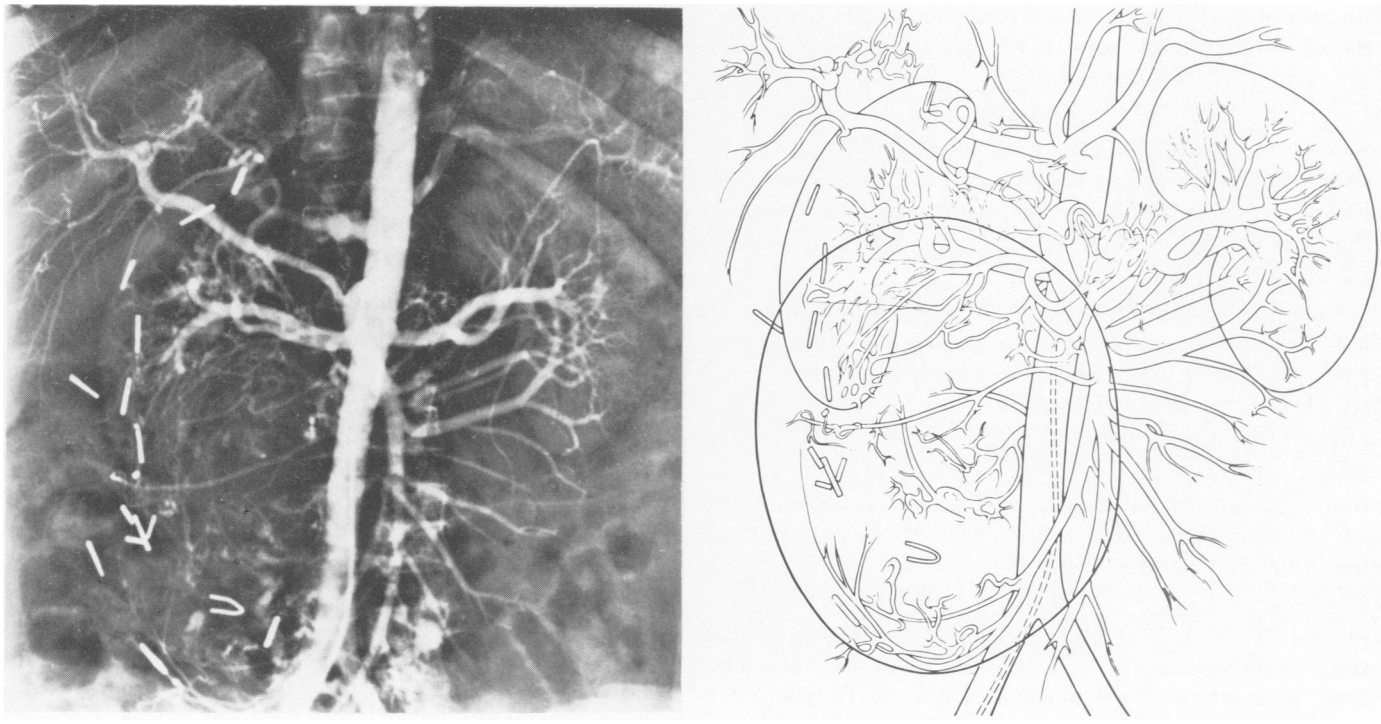
Author	Age	Sex	Operation	Diagnosis	Followup
Merkulow[39]	26	F	Attempted resection	Benign	Dead
Cahill[3]	10	F	Attempted	Benign	Dead
Haug & Baker[20]	60	M	Biopsy	Malignant	Dead, 6 mon postop
Holsti[26]	19	M	Attempted resection	Malignant	Well, 2 yr
Kinkhabwala[32]	—	—	Attempted resection	Malignant	—
Scharf[51]	33	F	Exploration	Malignant	Irradiation Dead, 11 yr postop

(Fig. 3). The appearance of this element is typical of non-chromaffin tumors of the paraganglia. Focally, dense bands of collagen were seen in all of the tumors, accounting for the lobular appearance (Fig. 4). In 3 of the tumors another cellular component was present. This consisted of sheets of cells with more pleomorphic nuclei and a more clearly defined granular, eosinophilic cytoplasm (Fig. 5), which were focally chromaffin-reacting as indicated by fine brown granules in specimen-fixed in Zenker's solution. The nuclei in some instances were bizarre and exhibited mitotic activity focally. These areas were also quite vascular and bands of dense fibrous tissue were also evident in some foci, but the nesting pattern was not present. The appearance of these areas was typical of pheochromocytoma. In a few areas admixtures of the 2 cell types were identified (Fig. 6). Metastasis to a regional lymph node was established histologically in only one instance.

Paragangliomas must be differentiated from carcinoid, hemangiopericytoma, alveolar soft part sarcoma and metastatic carcinoma, particularly thyroid carcinoma. The use of special histologic stains as an adjunct in differential diagnosis is summarized by Glenner and Grimley.[16] In cases of pheochromocytoma of the paraaortic area, a primary tumor in the adrenal medulla must be excluded.

Diagnosis

The symptoms of tumors arising in the organ of Zuckerkandl may be those related either to excessive



FIGS. 1a and b. (left) Aortogram visualizing the inferior mesenteric artery (Case 4). An abdominal aortogram shows a normal celiac axis. The opacified inferior mesenteric artery shows a large and very vascular tumor extending from just below the right renal artery to the level of the hypogastric artery. The right kidney is displaced slightly cephalad and its lower pole is distorted laterally. The tumor estimated to measure 20 cm in length and 10 cm in width rests to the right of the midline extending to the left of the aorta at the level of its greatest diameter.

The inferior mesenteric artery as it courses through the tumor is displaced to the left. It gives off many branches to both the substance and periphery of the mass. (right) Line drawing of aortogram.

catecholamine secretion or to the growth of a retroperitoneal mass.[45] Most of the patients, including all those seen at The New York Hospital, were seen because of excessive catecholamine secretion, and the symptoms in these cases are the same as in adrenal medullary pheochromocytomas. The clinical signs are likely to be headache, fainting and dizziness with hypertension, sometimes paroxysmal, in norepinephrine-secreting neoplasms. In epinephrine-secreting tumors the findings may be irritability, tachycardia, diarrhea, excessive sweating, flushing and possibly also hypertension. Many patients are aware that shift of position or pressure in an area within the abdomen may precipitate episodes. When the tumor arises in the organ of Zuckerkandl, the patient may be subject to paroxysms of hypertension following straining at stool or following ingestion of a large quantity of food or fluid. Palpation of the abdomen may reveal a tumor and pressure on the mass sometimes produces transient elevation of blood pressure.

In the evaluation of symptomatic patients, particularly those with hypertension, the most significant findings relate to elevation of urinary catecholamines and vanillyl mandelic acid (VMA) levels (total catecholamines normal range: 200–250 μg/24 hr; VMA normal range 8–9 μg/24 hr). Elevation of these substances during a period of hypertension as compared with those obtained in a resting period is of diagnostic significance. Blood values in patients with elevated urine determinations may also be of interest, but since those tests are expensive and time consuming, their routine use is not always practical. False positives, that is, patients with consistently elevated catecholamines and no demonstrable tumor, are documented but extremely rare. Provocative tests using histamine or regitine are dangerous and less specific than catecholamine determinations and are no longer a part of our diagnostic work-up.

In 80% of patients with elevated catecholamines, an adrenal tumor is likely to be found by intravenous pyelography. In absence of an adrenal lesion, chest roentgenograms may show a mediastinal tumor. Selective angiograms are most useful in demonstrating tumors arising from the organ of Zuckerkandl, and the evaluation of catecholamine levels of the inferior vena cava and its tributaries may also be of help. Although there has been reluctance in the past to use aortography in evaluation of patients with pheochromocytomas, this procedure is now widely accepted as essential and reasonably safe. Once the diagnosis is established, further confirmation may be obtained by use of blocking agents.

Preoperative Management

Adequate preoperative management is important in preventing the precipitous drop in blood pressure which may follow the release of the longstanding catecholamine vasospasm with sudden increase in vascular space. Preoperative alpha- and beta-adrenergic receptor blockades are invaluable in preoperative preparation. These substances are administered for days or weeks prior to operation, and, properly used, prevent cardiovascular disturbances during induction of anesthesia and the actual excision of the tumor. This is instituted with phenoxybenzamine hydrochloride 1 mg/kg orally and 45 mg of propranolol hydrochloride in 3 doses per day with gradual increases until the blood pressure and tachycardia are controlled. Patients who are well

FIG. 2. Functional paraganglioma of organ of Zuckerkandl (Case 4). This was a 244 gm lobulated, reddish-brown, retroperitoneal mass attached by fibrous bands and numerous vessels to the aorta, inferior mesenteric artery and inferior vena cava.

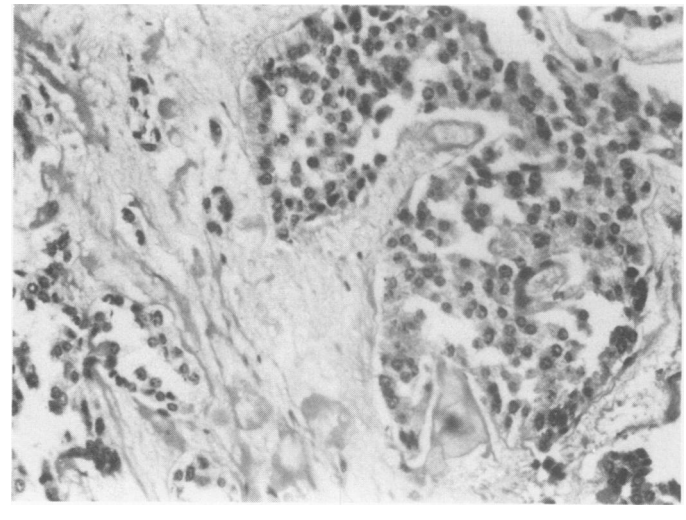


FIG. 3. Para-aortic paraganglioma of organ of Zuckerkandl. Alveolar nests of fairly uniform cells with round or oval vesicular nuclei and finely granular cytoplasm with indistinct borders are separated by thick bands of collagen. Cells of this type were the most prominent element in all 4 cases (H & E, ×62).

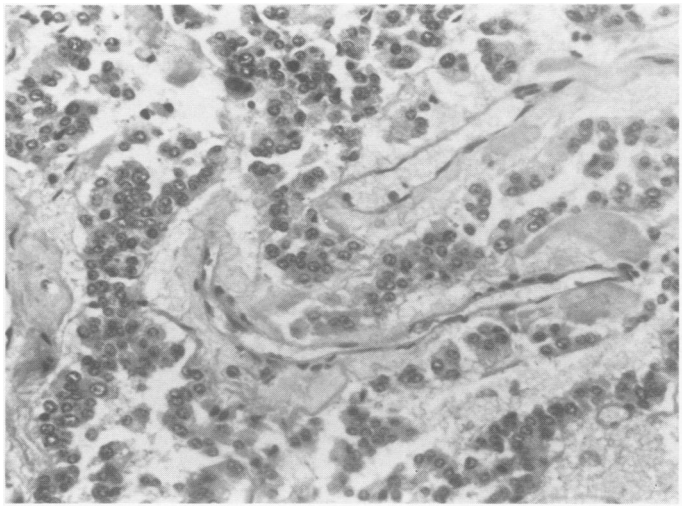


FIG. 4. Abundant blood vessels surrounded by dense collagen bands and separating nests of paraganglioma were seen in all of the tumors (H & E ×62).

prepared usually require less of a beta-blocker if it should be required at operation. The one death among our patients with a para-aortic paraganglioma occurred in 1962 prior to the availability of beta-blocker. Propranolol hydrochloride, which we now use, curtails ventricular arrhythmias that if uncorrected can result in death.

Preoperative medication also includes small doses of barbiturates and of atropine. With adequate preoperative preparation and suitable immediate preoperative medication and propranolol hydrochloride available in the operating room, induction of anesthesia is accomplished with intravenous pentothal. Venous cut-downs are placed so that norepinephrine or other pressor substances may be used at any time during the procedure if a fall of blood pressure occurs with the occlusion of the blood supply to the tumor. Continuous femoral artery pressure measurement by catheter is maintained throughout the procedure.

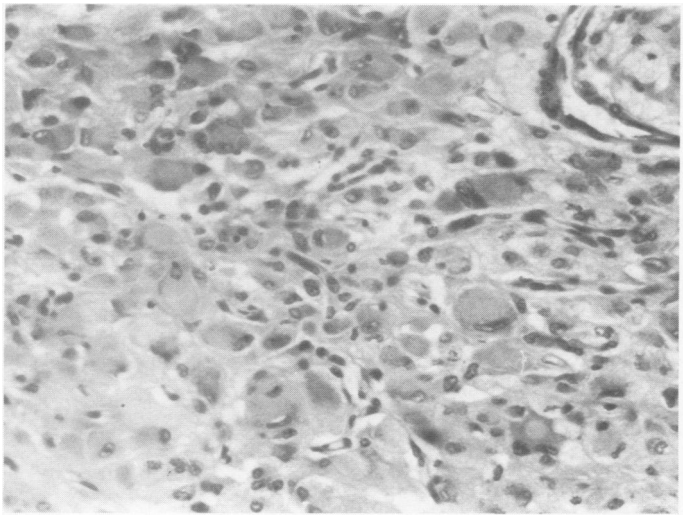


FIG. 5. Pheochromocytoma of organ of Zuckerkandl. Larger cells with distinct borders and granular cytoplasm grow in large sheets. Many of the nuclei are uniform and oval, but some larger, pleomorphic nuclei are seen along with multinucleated forms. Elements of this kind were identified focally in 3 cases (H & E ×62).

Surgical Technique

The surgical approach to tumors arising from the organs of Zuckerkandl is facilitated by generous incisions that provide optimum exposure of the lesion and adjacent structures.

Complete examination preliminary to attacking the tumor includes inspection and palpation of the adrenals and kidneys and demonstration of the course of the ureters from their origin to their junction with the urinary bladder. There may be displacement of these by the tumor if it is large. The second and third portions of the duodenum may also be distorted or displaced.

The relationship of the inferior mesenteric artery and its branches is to be anticipated and identified. The chief blood supply is usually from a number of vessels directly from the aorta. A comparable number of vessels extend directly from the posterior surface of the tumor to the inferior vena cava. These arterial and venous vessels tend to fix the tumor to the aorta and vena cava. Division of these vessels and the individual ligation of each is a tedious endeavor (Fig. 7).

The occlusion of the large vessels and their tributaries beyond the extent of the tumor and followed by division of the communicating vessels and then attempting to secure them as the occluding clamps are gradually released is even more difficult because of retraction of the small arterial branches into the wall of the aorta. These may not be apparent if the blood pressure falls to hypotensive levels. Then as the pressure returns to normal, bleeding tends to occur. During the operation of excising this type of tumor, occluding the return blood supply with its pressor substance contributes to lowering of blood pressures. Loss of blood may further accentuate the hypotension.

Discussion

Frankel[11] in 1886 was the first to postulate that a tumor of the adrenal was the cause of a malignant hypertension that resulted in the death of an 18-year-old girl upon whom he performed an autopsy. Mayo[36] presented the first report in English of a successfully treated case. In 1927 he operated upon a patient who had a large flank tumor, following excision of which there was a moderate, but not dramatic, fall in the blood pressure. It was possible to maintain the patient's vital signs by using intravenous saline, since at that time the pressor substances presently available were not in common use. Two years later, Pincoffs[47] made the preoperative diagnosis of a functioning paraganglioma in a

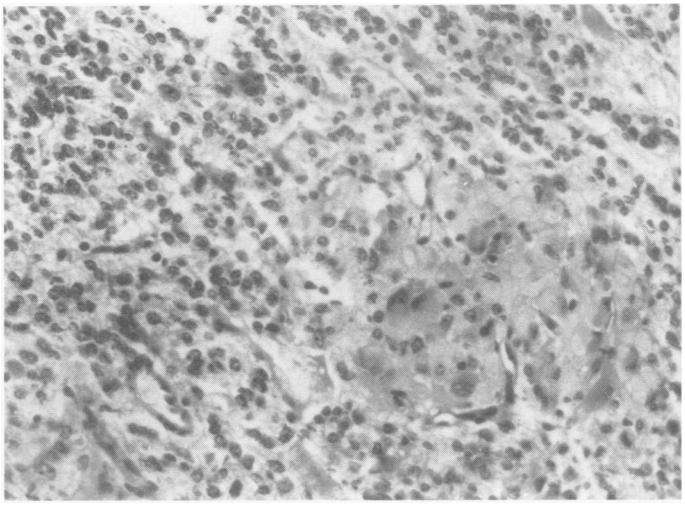


FIG. 6. Mixed cellular population in organ of Zuckerkandl tumor. A nodular cluster of pheochromocytoma type (lower right) is surrounded by paraganglion type cells. (H & E ×62.)

26-year-old woman who had repeated bouts of paroxysmal hypertension. After resection of the 115 gm right adrenal tumor by Shipley,[53] the blood pressure remained at very low levels but the patient recovered uneventfully and had a relatively normal life. Subsequently, further reports in the literature indicated that these tumors were being diagnosed with somewhat increasing frequency. The first adequate review was a collection of 207 cases published in 1951 by Graham.[17] At that time 125 patients had been subjected to surgery with 33 operative deaths. Seventy-two cases were not diagnosed before death, and it appeared that at least 44 of these patients died as a direct result of the hypertension. This would give an overall mortality up to 1951 of approximately 37%.

Between 1951 and 1960 many authors reported their experiences with pheochromocytomas. In 1960 Hume[27] collected over 700 cases from the world literature. He reported that as a result of the more frequent consideration of this diagnosis, the mortality rate was dropping steadily. Since 1960 several other large series of case reports have appeared in the English literature, the largest being the experience with over 100 patients at the Mayo Clinic published in 1966.[56]

Stackpole[54] in 1963 reviewed the experience of the Columbia-Presbyterian Medical Center and the world literature concerning this disease as it affected children. He also found that this disease was a more virulent and rapidly progressive one in children and that constant vigilance was necessary if the mortality rate was to be reduced. Many physicians, however, still fail to suspect pheochromocytoma as a cause for convulsion, elevation in blood pressure, fainting, intermittent blindness in children and therefore, it remains a particularly serious problem in this age group.

In the 1930's it became evident that many pheochromocytomas, probably at least 10%, were extra-adrenal, and that the most frequent site was in the region of the origin of the inferior mesenteric artery. Ortega[46] in 1952 published a report of a malignant paraganglioma arising from the organs of Zuckerkandl and summarized 14 reported tumors which were similarly located. Of 14 previously reported cases all but one were benign. The one malignant case could not be identified.

Retrospective studies then brought attention to Zuckerkandl's description in 1901 of paired bodies of extra-adrenal chromaffin cells near the origin of the inferior mesenteric artery. Bonnamour and Pinatelle[2] in 1902 confirmed Zuckerkandl's observations and described chromaffin bodies in children up to the age of 6 years. In 1912 Zuckerkandl[60] reviewed the literature relating to chromaffin tissues and the postnatal fate of these structures including tumor formation. Ivanoff[29] in 1925 de-

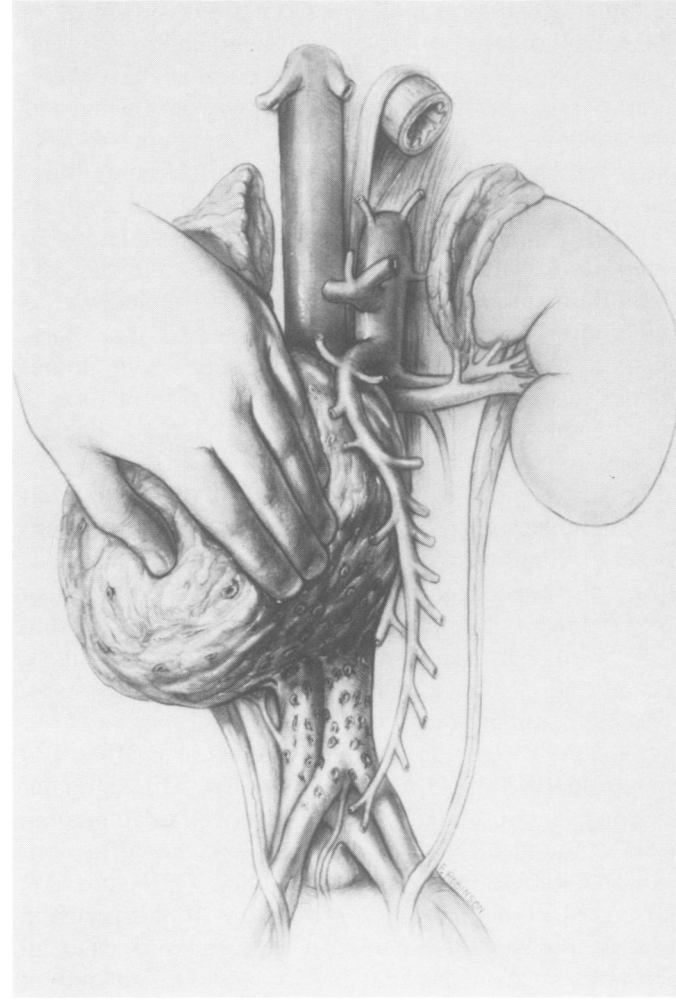


FIG. 7. At operation numerous short vessels were demonstrated to supply the tumor from the aorta. Comparable venous communications drained into the vena cava. These required individual division and ligation to mobilize the tumor, a tedious and time consuming endeavor.

scribed the development and postnatal degeneration of the paraganglia. Wrete[58] in 1927 observed the continuity of the chromaffin bodies of the para-aortic region with the chromaffin tissue of the adrenal glands. In the same year Keene and Hewer[31] observed the common origin of the intra- and extra-adrenal chromaffin cells from the sympathetic anlage.

The recent monograph of Glenner and Grimley[16] summarizes development of the concept of a system of paraganglia including the adrenal medulla and extra-adrenal chromaffin and non-chromaffin tissues. All paraganglia develop from neural crest and migrate with elements of the autonomic nervous system to para-axial locations in association with autonomic ganglia. The paraganglia are more prominent in infancy and, except for the adrenal medulla, are inconspicuous in adults. Branchiomeric paraganglia, including the carotid bodies and glomus jugulare, appear to function primarily as chemoreceptors. Aortico-sympathetic paraganglia, including the adrenal medulla and para-aortic ganglia (the organ of Zuckerkandl) are specialized for secretion of catecholamines, although catecholamine storage granules are demonstrable in all paraganglia. Many instances are reported of tumors arising from the carotid body and glomus jugulare, and paraganglionic tumors have been documented in larynx, mediastinum, lung, orbit, vagus and bladder. Those of the adrenal medulla and organ of Zuckerkandl often have functional activity related to excessive secretion of pressor substances. In addition, tumors of other paraganglia, including the branchiomeric group, occasionally secrete catecholamines in quantities sufficient to produce symptoms. Glenner *et al.*[15] documented functional carotid tumors and Mitsuguahi *et al.*[41] have reviewed functional jugular paraganglioma (glomus jugulare tumor).

The histologic pattern of para-aortic (Zuckerkandl) paragangliomas varies and may be either as the non-chromaffin, paraganglioma (carotid body) type or as chromaffin pheochromocytoma type or as mixtures of these elements. The paraganglioma pattern was present in all 4 of our cases and was the most prominent type in the 3 cases with mixed histologic patterns. Histologic appearance is inconsistently related to catecholamine secretion since both chromaffin and non-chromaffin tumors may be functional. Although non-functional, non-chromaffin tumors are well documented, we have found no well established instances of a non-functional, para-aortic chromaffin tumor. Unfortunately, assessment of histologic type in many of the reported cases is impossible. Histologic appearance is also an unreliable measure of malignancy and benignancy of Zuckerkandl tumors. Pleomorphism, bizarre nuclear forms and mitotic figures are common findings in pheochromocytomas and do not necessarily connote metastatic potential. Likewise, nuclear uniformity and circumscription may be seen in non-chromaffin tumors with nodal metastases. The only reliable criteria for diagnosis of malignancy is demonstration of invasion of adjacent organs or of metastases. Because of the dense adhesions between many of these tumors and surrounding tissues, assessment of invasion is often difficult. Even nodal metastases do not necessarily indicate an aggressive tumor, as indicated by the long term survival of one of our patients (Case 1), with tumor found in a lymph node nearby the primary tumor. Nevertheless, the finding of tumor within a lymph node indicates the malignant potential of some of these tumors.

An estimated 22,950,000 individuals in the United States have hypertension.[22] In a study at the Cleveland Clinic 6% of 5000 patients were demonstrated to have a cause that could be successfully treated surgically.[14] Of these, functioning pheochromocytoma is one of the most favorable for the surgeon to approach, since removal of those tumors that are benign results in a cure. However, 10–15% of catecholamine secreting neoplasms are extra-adrenal, and the largest number of these arise from the organ of Zuckerkandl.

The relative young ages of the patients we have studied and of those reported by others indicates that hypertension in children and young adults merits most meticulous evaluation to permit diagnosis and treatment of a functional paraganglioma before development of irreversible sequelae.

Addendum

Since this paper was submitted, Patient 4, 20 months following his first operation has had an epidural metastatic paraganglioma removed from the L4-5 interspace. The pathological specimen is identical with the primary tumor.

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Discussion

(Note: Some of the discussants' remarks refer to both this paper and to the one following by Dr. H. William Scott, Jr.)

Dr. William H. ReMine (Rochester, Minnesota): When we reviewed our experience before this group two years ago, we had at that time 14 extra-adrenal pheochromocytomas, [slide] and as this has been known to be the 10% tumor, ours came out to be right on the button on 10% of the total group, as you can see.

In reviewing our extra-adrenal pheochromocytomas, this was the distribution as we found them to be at that time. Since this paper was reported, and these figures accumulated, we have had additional cases of this type. One was in the glomus jugulari at the base of the skull. As you know, chromaffin tissue and paraganglia tissue can occur from the base of the skull all the way to a point deep in the